PART XXII Cancer and Benign Tumors

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1. Which of the following malignant tumors is least likely to occur in adults in comparison with children?

A. acute Lymphoblastic Leukemia (ALL)
B. osteosarcoma
C. medulloblastoma
D. retinoblastoma
E. Hodgkin lymphoma

2. A 9-year-old boy develops acute myelogenous leukemia (AML) one year after completion of therapy for soft tissue sarcoma at his right thigh.
Which of the following chemotherapeutic agents is **MOST** likely the cause of secondary acute myelogenous leukemia AML in this boy?

A. cyclophosphamide
B. vincristine
C. etoposide
D. doxorubicin
E. cisplatin

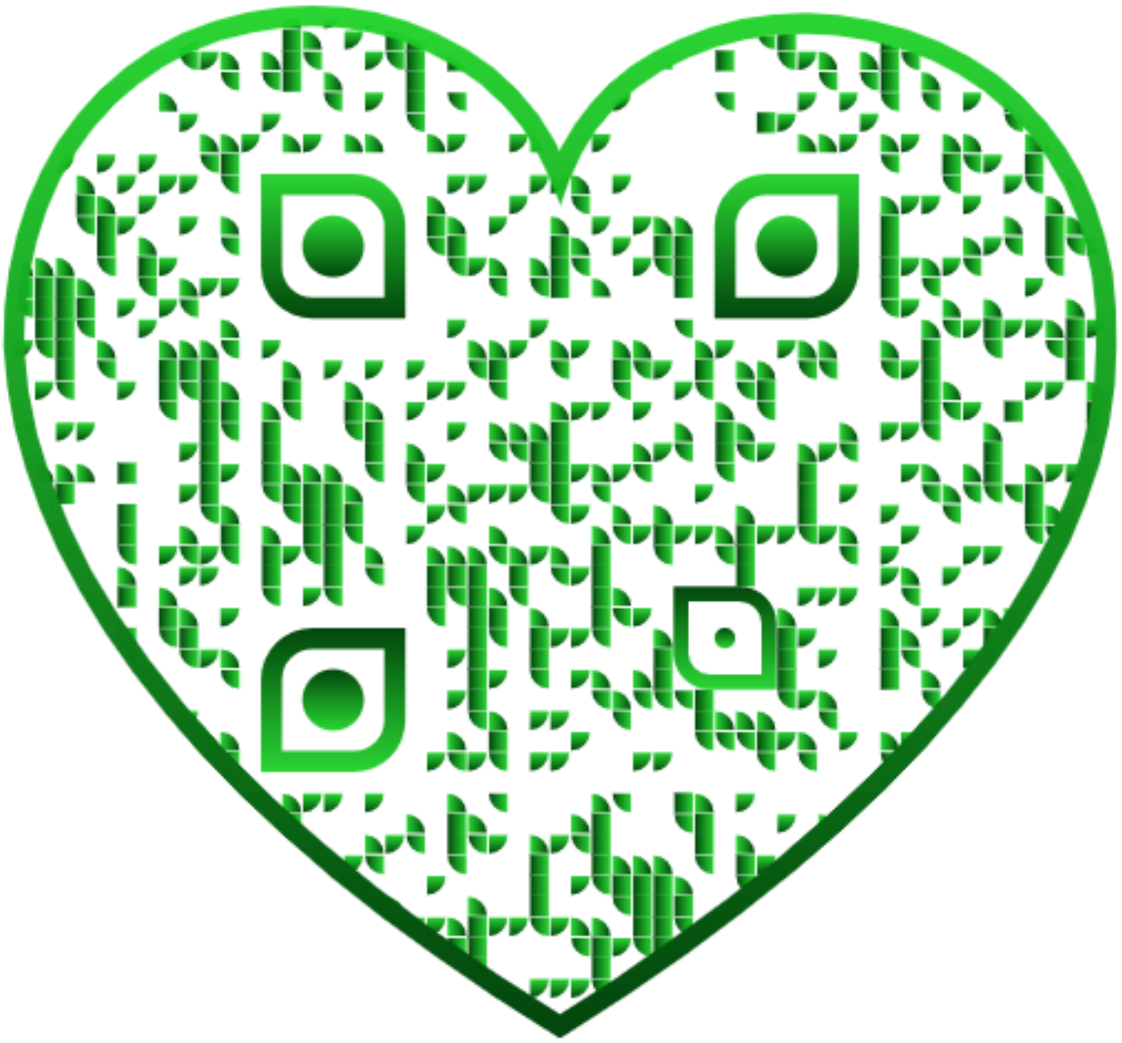
3. Of the following, the genetic syndrome **MOST** likely associated with increased risk of optic glioma is

A. Down syndrome
B. neurofibromatosis 1
C. monosomy 7
D. Bloom's syndrome
E. ataxia-telangiectasia

4. Constellation of aniridia and hemihypertrophy is strongly associated with increased risk of which of the following tumors?

A. rhabdomyosarcoma
B. hepatoblastoma
C. Wilms` tumor
D. neuroblastoma
E. medulloblastoma

5. During the routine exam of an infant, the parents state a strong family history of adenomatous polyposis.



The statement that should be included during the discussion is

A. the infant is at increased risk for colonic adenocarcinoma
B. the infant is at increased risk of acute lymphocytic leukemia
C. the infant is at increased risk of intestinal Burkitt lymphoma
D. the infant is at increased risk of hepatoblastoma
E. the infant is at increased risk of germ cell tumor

6. Undescended testis is a risk factor for the development of which of the following tumors?

A. rhabdomyosarcoma
B. leukemia
C. yolk sac tumor
D. lymphoma
E. neuroblastoma

7. Epstein-Barr virus (EBV) infection is more likely to be associated with all the following malignancies **EXCEPT**

A. Burkitt lymphoma
B. nasopharyngeal carcinoma
C. T-cell lymphoma
D. Hodgkin lymphoma
E. hepatocellular carcinoma

8. Which of the following malignancies is least likely to occur in a 10-month-old infant?

A. neuroblastoma
B. nephroblastoma
C. retinoblastoma
D. hepatoblastoma
E. Ewing sarcoma

9. Hepatitis C virus infection is a risk factor for which of the following malignancy?

A. hepatoblastoma
B. splenic lymphoma
C. Hodgkin lymphoma
D. nasopharyngeal carcinoma
E. cervical carcinoma

10. Nearly all cervical carcinomas contain human papillomavirus (HPV). Which of the following types is never associated with such malignancy?

A. type 6
B. type 16
C. type 18

D. type 31
E. type 33

11. Which of the following chromosomal abnormalities in acute lymphoblastic leukemia of childhood carry a favorable outcome?

A. t(12;21)
B. t(4;11)
C. t(9;22)
D. hypodiploidy
E. 11q23

12. You are meeting with parents of 10-year-old child who recently develops acute lymphoblastic leukemia (ALL); the mother has a concern about risk of CNS relapse.
Which of the following is **LEAST** likely to increase the risk of CNS relapse in children with ALL?

A. first traumatic lumbar puncture (LP)
B. T-cell leukemia
C. cranial nerve involvement at the time of diagnosis
D. presence of lymphoblast in the CSF at any time during treatment
E. age of more than 10 year at the time of the diagnosis

13. Children with ALL who carry poor outcome include all the following **EXCEPT**

A. age younger than 1 year and older than 10 year
B. T-cell immunophenotype
C. hyperdiploidy chromosomal abnormality
D. initial leukocyte count of > 50,000
E. poor response to initial therapy

14. Which of the following chromosomal abnormalities of childhood ALL carries the highest risk of relapse despite intensive chemotherapy?

A. t(9;22)
B. t(4;11)
C. hypodiploidy
D. t(1;19)
E. t(12;21)

15. You are meeting the parents of a child with ALL who want to go for a trip with their child. The **MOST** nontoxic phase of the treatment of ALL that allow the family to have a safe trip is

A. before starting treatment
B. remission induction
C. consolidation

D. delayed intensification
E. interim maintenance

16. You are evaluating a 6-year-old child with ALL on interim maintenance phase who has frequent mucositis and myelosuppression that needs frequent discontinuation of his treatment.
Of the following, the **MOST** valuable test for this child is
A. complete blood count
B. bone marrow study
C. pharmacogenetic testing of the thiopurine s-methyltransferase (tpmt) gene
D. lumbar puncture
E. renal function test

17. You are evaluating a 9-year-old boy child with ALL who recently develops testicular relapse; an important statement that should be mentioned to his parents is
A. testicular relapse occurs in the majority of boys with ALL
B. it usually occurs during the course of therapy
C. such relapse occurs as painful swelling of oneor both testes
D. the diagnosis is confirmed by ultrasonography of the affected testis
E. the majority of affected boys can be successfully retreated, and the survival rate is good

18. Granulocytic sarcoma masses are commonly seen in which type of the following malignancies?
A. neuroblastoma
B. rhabdomyosarcoma
C. soft tissue sarcoma
D. ALL
E. AML

19. Which of the following types of translocation of childhood AML that typically associated with granulocytic sarcoma mass?
A. t(15:17)
B. t(8:21)
C. inv(16)
D. t(6:9)
E. inv(3)

20. You are explaining the risk of leukemia in children with Down syndrome to medical students; your discussion will include all the following statements **EXCEPT**
A. acute leukemia occurs more frequently in children with Down syndrome than in the general population

B. AML is more common in children with Down syndrome in the 1st 3 yr of life than ALL as compared with the ratio of ALL/AML in general population
C. in children with Down syndrome who have AML, the expected outcome of treatment is slightly inferior to that for other children
D. patients with Down syndrome who develops acute leukemia demonstrate a remarkable sensitivity to antimetabolites
E. neonates with Down syndrome may develop a transient leukemia or myeloproliferative disorder

21. A healthy 20-day-old male neonate with Down syndrome appears pale; examination reveals a palpable liver 6 cm below the right costal margin and palpable spleen 3 cm below the left costal margin; lab findings include: hemoglobin, 8.8 g/dl; platelets count, 55000/mm3; white blood cell count, 18700/mm3 with 10% blast cells; bone marrow examination is consistent with acute leukemia.
Of the following, the **BEST** approach for the management is
A. intensive chemotherapy
B. low dose chemotherapy
C. pulses of chemotherapy
D. bone marrow transplantation
E. close follow up

22. Which of the following types of leukemia is **MOST** likely to develop in a 2-year-old child with Down syndrome who has been developed transient myeloproliferative disorder in the neonatal period?
A. ALL
B. CML
C. AML M1
D. AML M6
E. AML M7

23. Which of the following chromosomal abnormalities is often characteristic of infantile ALL?
A. t(4;11)
B. t(12;21)
C. t(9;22)
D. t(10;14)
E. t(1;19)

24. Hodgkin lymphoma in children usually has favorable outcome; however, the prognosis may be grim in some cases.
Poor prognostic factors in Hodgkin Lymphoma include all the following **EXCEPT**
A. age of more than 15 year at the time of the diagnosis

B. stage IV disease
C. presence of “B” symptoms
D. poor response to therapy manifested by positron emission tomography (PET) scan positivity
E. presence of bulky mediastinal mass

25. A 12-year-old male adolescent, recently diagnosed with Hodgkin lymphoma; the PET-CT scan shows an involvement of the left cervical and supraclavicular lymph nodes; he has a history of drenching night sweating and high grade fever.
Based on Ann Arbor Staging Classification for Hodgkin Lymphoma, the patient is classified as

A. stage IA
B. stage IB
C. stage IIA
D. stage IIB
E. stage IIIB

26. Which of the following manifestations is more specific to occur in anaplastic large cell lymphoma (ALCL) than other types of non-Hodgkin Lymphoma (NHL)?

A. primary intestinal involvement
B. bone marrow involvement
C. CNS involvement
D. primary cutaneous involvement
E. primary mediastinal involvement

27. Tumor lysis syndrome (TLS) is a common complication during the treatment of pediatric malignancies, which of the following malignancies is more likely to manifest such complication?

A. acute myelogenous leukemia AML
B. nephroblastoma
C. neuroblastoma
D. Burkitt lymphoma
E. Rhabdomyosarcoma

28. Metabolic derangement secondary to tumor lysis syndrome is commonly encountered after starting chemotherapy in children with malignancy; it include all the following **EXCEPT**

A. hyperuricemia
B. hypernatremia
C. hyperkalemia
D. hyperphosphatemia
E. hypocalcemia

29. A 5-year-old child, recently diagnosed with ileocecal Burkitt lymphoma; lab investigations reveal: serum uric acid 12 mg/dl, serum sodium, 145 meq/dl; serum potassium, 4.5 meq/dl; serum phosphate 4.4 meq/dl; serum calcium, 8.9 mg/dl; blood urea, 22 mg/dl; serum creatinine, 0.8 mg/dl.
Of the following, the **MOST** effective treatment is

A. excessive hydration
B. sodium bicarbonate
C. xanthine oxidase inhibitor
D. recombinant urate oxidase
E. no treatment

30. A 6-year-old male child presented with a cerebellar mass; after undergoing complete surgical resection of the mass, the histology of the mass reveals pilocytic astrocytoma.
Of the following, the **MOST** appropriate next step in the management is

A. radiotherapy
B. chemotherapy
C. observation
D. concomitant chemo-radiotherapy
E. radiotherapy followed by chemotherapy

31. A 6-year-old child complains of bilateral thigh pain, motor weakness and some sensory deficits of both lower limbs; he has a history of bladder dysfunction over the past 2 weeks; MRI of the spinal cord shows a mass arising from the filum terminale and conus medullaris causing some pressure effect. A CNS tumor is suspected.
Which of the following tumor is **MOST** likely arising in such site?

A. medulloblastoma
B. anaplastic astrocytoma
C. myxopapillary ependymoma
D. choroid plexus carcinoma
E. oligodendroglioma

32. You are discussing the risk of radiotherapy with the parents of a child with medulloblastoma; the mother has a concern about the late neurological complications post radiotherapy.
The statement that should be included in the discussion that late neurological sequelae post radiotherapy is more severe with

A. focal radiotherapy rather than craniospinal irradiation
B. children with an age of less than 3 year
C. concomitant chemo-radiotherapy
D. low grade tumors rather than high grade tumors
E. infratentorial tumors rather than supratentorial tumors

33. The least common late neurologic sequelae that may be encountered post craniospinal irradiation for a 9-year-old child with medulloblastoma is

A. microcephaly
B. learning disabilities
C. cognitive impairment
D. neuroendocrine dysfunction
E. second malignancy

34. Chemotherapy has a major role in many childhood CNS tumors. In which of the following tumors chemotherapy is not effective?

A. medulloblastoma
B. pilocytic astrocytoma
C. craniopharyngioma
D. pineoblastoma
E. supratentorial primitive neuroectodermal tumors (SPNETs)

35. Childhood primary brain stem tumors are heterogenous group of tumors; the outcome usually depends on tumor location.
Which of the following brain stem tumors, depending on the site of tumor, carry the worst prognosis?

A. focal
B. dorsally exophytic
C. cervicomedullary
D. diffuse intrinsic
E. none of the above

36. Extraneural metastasis from primary brain tumors is less likely to occur. Which of the following primary brain tumor is **MOST** commonly metastasizing extraneurally?

A. medulloblastoma
B. primitive neuroectodermal tumor (PNET)
C. ependymoma
D. malignant glioma
E. choroid plexus carcinoma

37. Childhood primary brain tumors are less likely to metastasize extraneurally. Which of the following is **MOST** likely considered a risk factor for extraneural metastasis?

A. supratentorial tumor
B. age of the patient of less than 10 year
C. female gender
D. ventriculoperitoneal (VP) shunt insertion
E. glial tumors

38. Small round blue cell tumors refer to tumors that have round blue cells histologically. They include all the following **EXCEPT**

A. neuroblastoma
B. non-Hodgkin lymphoma
C. osteosarcoma
D. Ewing sarcoma
E. rhabdomyosarcoma

39. A 9-mo-old infant develops a left adrenal mass; histological examination reveals a significant degree of undifferentiated round blue cells consistent with neuroblastoma.
Which of the following genetic characteristics of the tumor cells carry a better outcome?

A. amplification of the MYCN (N-myc) proto-oncogene
B. hyperdiploidy
C. loss of heterozygosity of 1p chromosome
D. loss of heterozygosity of 11q chromosome
E. gain of 17q chromosome

40. A 1-year-old child develops right eye ptosis, myosis, and loss of sweating; you suspect neuroblastoma.
Of the following, the **MOST** valuable investigation to confirm the diagnosis is

A. Computerized tomography (CT) scan of the neck and chest
B. CT scan of the abdomen
C. CT scan of the brain
D. magnetic resonance imaging (MRI) of the brain
E. X-ray the chest

41. Neuroblastoma is a highly malignant childhood neoplasm. Metastatic spread can occur via local invasion or distant hematogenous or lymphatic routes.
Of the following, the **LEAST** common site of metastases in neuroblastoma is

A. long bones
B. bone marrow
C. lung
D. skin
E. liver

42. Neuroblastoma can be associated with paraneoplastic syndrome. All the following are paraneoplastic features of neuroblastoma **EXCEPT**

A. uncontrollable jerking eye and body movements
B. cerebellar ataxia and poor coordination
C. increased sweating and hypertension
D. unilateral ptosis, myosis, and anhidrosis

E. profound secretory diarrhea

43. You are evaluating a 6-mo-old girl with a firm abdominal mass. Radiography reveals a calcified small right suprarenal mass. The tumor is completely resected and histologically is confirmed as neuroblastoma. Metastatic work-up shows massive liver involvement, 10% bone marrow involvement by tumor cells, and multiple subcutaneous nodules involvement. There is no bony involvement. The *N*-myc oncogene from the tumor is not amplified.
According to the international neuroblastoma staging system, the infant is stratified as

A. stage I
B. stage II A
C. stage III
D. stage IV
E. stage IV S

44. A 10-mo-old boy has a left suprarenal mass. Surgery is accomplished with complete surgical removal of the mass as well as the non-adherent ipsilateral lymph nodes; multiple contralateral lymph nodes biopsies are taken during surgery. The histology reveals poorly differentiated neuroblastoma with microscopic involvement of the ipsilateral lymph nodes; the contralateral lymph nodes are negative for tumor cells.
Of the following, the **BEST** therapeutic approach for this infant is

A. chemotherapy
B. radiotherapy
C. concomitant chemo-radiotherapy
D. chemotherapy followed by radiotherapy
E. close observation

45. Children with high-risk neuroblastoma have poor survival. Current treatment consists of all the following E**XCEPT**

A. surgery
B. intensive chemotherapy
C. radiation
D. allogenic bone marrow transplantation
E. differentiating agents (13-cis-retinoic acid)

46. A 2-year-old male child has an asymptomatic right flank mass discovered incidentally by the mother while bathing. Suspicion of Wilms` tumor is raised.
Of the following, the **LEAST** likely investigation to be performed in this child is

A. plain abdominal radiography
B. computerized tomography (CT) scan of the abdomen
C. CT scan of the chest
D. Biopsy of the mass

E. ultrasonography with Doppler imaging of the abdominal blood vessels

47. Biopsy for children with Wilms` tumor is not usually performed as it may upstage the disease.
All the following are indications of accomplishing renal biopsy for a child with suspicion of Wilms` tumor **EXCEPT**

A. age of 2-3 year at presentation
B. signs of infection and inflammation around the tumor
C. significant abdominal lymph nodes enlargement radiologically
D. absence of renal parenchyma radiologically
E. radiological appearance of intratumoral calcification

48. A 2-year-old child is being evaluated for a right flank mass; radiological appearance is consistent with right renal Wilms` tumor. During radical nephrectomy there is intra-abdominal tumor rupture stated by the surgeon.
Of the following, the **BEST** therapeutic approach for this child is

A. chemotherapy
B. radiotherapy
C. concomitant chemotherapy-radiotherapy
D. another surgery
E. observation

49. Although children with Wilms` tumor have a favorable prognosis, there are some adverse factors that affect outcome.
All the following are poor prognostic factors in children with Wilms` tumor **EXCEPT**

A. young age at diagnosis
B. advanced stage
C. large tumors
D. anaplastic histology
E. loss of heterozygosity at chromosome 1p and 16q

50. Although it is not common, about 15% of children with Wilms` tumor suffer relapse, most relapses occur early (within 2 yr of diagnosis).
Factors associated with a favorable outcome after relapse include all the following **EXCEPT**

A. low stage (I/II) at diagnosis
B. no prior radiotherapy
C. anaplastic histology
D. relapse to lung only
E. late relapse more than 12 mo from nephrectomy

51. A 2-mo-old male infant has a left flank mass discovered incidentally by the mother; radiological imaging reveals a left renal mass.
Of the following, the **MOST** likely diagnosis is

A. nephroblastoma
B. neuroblastoma
C. mesoblastic nephroma
D. clear cell sarcoma
E. rhabdoid tumor of the kidney

52. A healthy 4-year-old child has a left flank mass. Computerized tomography (CT) scan of the abdomen shows a localized left renal mass. Radical nephrectomy is performed; the histology shows clear cell sarcoma of the kidney.
Of the following, the **MOST** appropriate next step in the management is

A. CT scan of the chest
B. CT scan of the brain
C. bone scan
D. magnetic resonance imaging (mri) of the brain
E. skeletal survey

53. Renal cell carcinoma (RCC) is rare in children, accounting for <5% of all renal tumors of childhood.
All the following are true regarding RCC in children **EXCEPT**

A. patients may present with frank hematuria, flank pain, and/or a palpable mass
B. it can be asymptomatic and detected incidentally
C. it has a propensity to metastasize to the lungs, bone, liver, and brain
D. it can be associated with von hippel–lindau disease
E. local lymph node involvement has an adverse outcome

54. Alveolar type of rhabdomyosarcoma (RMS) accounts for approximately 1/3 of all cases of pediatric RMS and carries the poorest prognosis.
Of the following, the **MOST** common site of involvement by alveolar type RMS is

A. orbit
B. middle ear
C. extremities
D. vagina
E. bladder

55. In pediatric rhabdomyosarcoma, stages are dependent on primary site whether favorable or unfavorable.
Which of the following sites of involvement is considered unfavorable?

A. vagina
B. uterus

C. testis
D. orbit
E. paranasal sinuses

56. Osteosarcoma is the most common primary malignant bone tumor in children and adolescents; it has multiple subtypes and requires different modalities of treatment including surgery and chemotherapy.
Of the following, the subtype of osteosarcoma which is treated by surgery alone is
A. fibroblastic
B. chondroblastic
C. telangiectatic
D. parosteal
E. periosteal

57. Of the following, the **WORST** prognostic factor in pediatric osteosarcoma is
A. primary pelvic bone tumor
B. poor histologic response to treatment
C. bony metastases at the time of diagnosis
D. lung metastases at the time of diagnosis
E. chondroblastic subtype

58. Radiotherapy is an effective modality of treatment in variable pediatric solid tumors.
Of the following, the **LEAST** responsive tumor to radiotherapy is
A. rhabdomyosarcoma
B. neuroblastoma
C. nephroblastoma
D. osteosarcoma
E. Ewing sarcoma

59. A 12-year-old male adolescent has pain of the right upper thigh that gradually becomes more severe and most often at night; it usually relieved by taking salicylates medication. Examination reveals limping, atrophy, and weakness of the right lower extremity. Plain radiography shows a round lucent lesion at the diaphysis of the right upper femur, about 0.5 cm in diameter, surrounded by sclerotic cortical bone formation.
Of the following, the **MOST** likely diagnosis is
A. Ewing sarcoma
B. osteosarcoma
C. osteoid osteoma
D. osteoblastoma
E. nonossifying fibroma

60. During a routine examination of a 10-mo-old male infant, you find a white pupillary reflex of right eye; the eye movements are normal. You suspect retinoblastoma.
Of the following, the **BEST** confirmatory diagnostic evaluation of this infant is

A. indirect ophthalmoscopy with slit-lamp examination
B. examination under general anesthesia by an experienced ophthalmologist
C. retinal biopsy
D. orbital ultrasonography
E. brain MRI

61. A 9-mo-old infant is recently diagnosed with bilateral retinoblastoma; examination under anesthesia reveals bilateral multifocal involvement of the retina. An important step in the management is

A. radiotherapy of both eyes
B. retinal examination of 1st degree relatives
C. orbital ultrasonography
D. bilateral bone marrow examination
E. bilateral enucleation

62. The serum α-fetoprotein (AFP) level is elevated with some malignant germ cell tumors (GCTs) especially endodermal sinus tumors; it can be used as a measure of response to treatment and during follow up after completion of chemotherapy. However, it is normally elevated during infancy.
At which age does AFP physiologically fall to normal adult level?

A. three month
B. eight month
C. one year
D. three year
E. ten year

63. The β subunit of human chorionic gonadotropin (HCG) is a useful surface marker in some malignant germ cell tumors (GCTs); it is secreted by syncytiotrophoblasts.
In which of following GCTs this surface marker is characteristically elevated?

A. teratoma
B. endodermal sinus tumor
C. germinoma
D. gonadoblastoma
E. embryonal carcinoma

64. You are discussing with medical students the role of chemotherapy in malignant germ cell tumors (GCTs); you state that GCTs are sensitive to some types of chemotherapy.
Of the following, the **MOST** effective chemotherapeutic agent in GCTs is

A. vincristine
B. cyclophosphamide
C. cisplatin
D. methotrexate
E. cytosine Arabinoside

65. Hepatoblastoma occurs predominantly in children younger than 3 yr of age. Although etiology is unknown, there are many associated risk factors.
All the following are associated risk factors for development of hepatoblastoma **EXCEPT**
A. familial adenomatous polyposis
B. Beckwith-Wiedemann syndrome
C. prematurity
D. low birth weight
E. hepatitis C infection

66. Hepatoblastoma is the most common neoplasm of the liver; it arises from the precursors of hepatocytes. It is of different histological classification which has a direct correlation with clinical outcome.
Of the following, the histological type that predict the **MOST** favorable outcome is
A. epithelial type of pure fetal histology
B. epithelial type of pure embryonal histology
C. epithelial type of mixed histology (fetal and embryonal)
D. mixed type of epithelial and mesenchymal elements
E. undifferentiated type

67. Hemangiomas are the most common benign tumors of infancy, occurring more often in the head and neck region.
Of the following, the infant at higher risk of development of hemangioma is
A. full term infant
B. male gender infant
C. infant of woman who had chorionic villus sampling
D. infant delivered by cesarean section
E. infant of diabetic mother

68. You are examining an infant with multiple cutaneous hemangiomas; you suspect an involvement of internal organs.
Of the following, the **MOST** common site of visceral involvement by hemangiomas is
A. brain
B. heart
C. lung
D. liver
E. pancreas

69. Kasabach-Merritt syndrome is a benign vascular tumor which may be life-threatening; it is characterized by all the following **EXCEPT**

A. rapidly enlarging lesion
B. thrombocytopenia
C. microangiopathic hemolytic anemia
D. coagulopathy
E. association with infantile hemangiomas

70. A 3-day-old neonate has a large soft, painless mass involving the right lower part of the head and neck that transilluminate; computerized tomography reveals a cystic mass infiltrating the neck and intrathoracic region mostly the mediastinum.
Of the following, the **BEST** modality of treatment for this neonate is

A. aspiration of mass
B. surgical resection
C. Injection of sclerosing agent
D. laser therapy
E. systemic interferon

71. You are meeting with parents of 12-year-old girl who recently diagnosed with papillary thyroid carcinoma (PTC). The statement that should be included in your discussion is

A. PTC has a grim overall prognosis
B. chemotherapy is the primary therapy
C. children with PTC do not require radioactive iodine therapy (^{131}I)
D. supraphysiologic levothyroxine is required during the course of treatment
E. long-term follow-up involves monitoring of calcitonin/carcinogenic embryonic antigen tumor markers

72. You are examining a 12-year-old female adolescent with a small nevus in the left thigh; her mother is concerned regarding development of melanoma in future.
All the following findings raise the suspicion of melanoma **EXCEPT**

A. rapidly enlarging nevus
B. nevus with changing colors
C. nevus with irregular margins
D. easily bleeds nevus
E. Spitz nevus

73. Although melanoma is relatively rare in children, some risk factors may increase its incidence.
All the following are risk factors for development of melanoma **EXCEPT**

A. positive family history of melanoma
B. dark skinned child

C. giant hairy nevus
D. dysplastic nevus syndrome
E. xeroderma pigmentosum

74. Nasopharyngeal carcinoma is one of the most common nasopharyngeal tumors in pediatric patients. Many factors may affect the prognosis.
Which of the following carries the worst outcome?
A. elevated lactate dehydrogenase (LDH) level
B. advanced local disease
C. extensive cervical lymph nodes involvement
D. Epstein-Barr virus (EBV) DNA levels
E. evidence of distant metastases

75. You are evaluating a 6-year-old male child with Langerhans cell histiocytosis (LCH); you states to the parents that skeletal survey and bone scan are important investigations as bones are the most common sites of involvement.
Of the following, the **MOST** common site of bone involvement by LCH is
A. skull
B. mandible
C. vertebra
D. pelvis
E. femur

76. Langerhans cell histiocytosis (LCH) has an extremely variable presentation. Single or multiple organs may be affected by the disease.
Of the following, the **LEAST** likely organ affected by LCH is
A. skin
B. bone
C. middle ear
D. lung
E. lymph nodes

77. In Langerhans cell histiocytosis (LCH), systemic manifestations are more likely to occur in patients with multisystem disease who are at high risk of mortality (i.e., "risk organ"-positive patients). All the following are risk organs in LCH **EXCEPT**
A. bone
B. liver
C. spleen
D. lung
E. hematopoietic system

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PART XXII Cancer and Benign Tumors ANSWERS

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1.**(D).** Retinoblastoma is an embryonal tumor, usually occurs in childhood.
2.**(C).** Epipodophyllotoxins like etoposide may cause secondary AML early within two years after administration; alkylating agents like cyclophosphamide may cause secondary AML as far as 5 years after therapy.
3.**(B).** Neurofibromatosis 1 is strongly associated with optic glioma. Down syndrome and Monosomy 7 are associated with increased risk of AML. Bloom's syndrome and ataxia-telangiectasia are associated with increased risk of ALL.
4.**(C).** Beckwith-Wiedemann syndrome, an overgrowth syndrome characterized by macrosomia, macroglossia, hemihypertrophy, omphalocele, and renal anomalies, is associated with an increased risk of Wilms` tumor, hepatoblastoma, rhabdomyosarcoma, neuroblastoma, and adrenal cortical carcinoma, while aniridia is associated with Wilms` tumor.
5.**(D).** Beckwith-Wiedemann syndrome, hemihypertrophy, Gardner syndrome, and family history of adenomatous polyposis are associated with increased risk of hepatoblastoma.
6.**(C).**
7.**(E).** Hepatocellular carcinoma is associated with hepatitis B and C infection.
8.**(E).** During the 1st yr of life, embryonal tumors such as neuroblastoma, nephroblastoma (Wilms tumor), retinoblastoma, rhabdomyosarcoma, hepatoblastoma, and medulloblastoma are most common. As children age, bone malignancies, Hodgkin disease, gonadal germ cell malignancies (testicular and ovarian carcinomas), and other carcinomas increase in incidence.
9.**(B).** Hepatitis C virus infection is a risk factor for hepatocellular carcinoma and is also associated with splenic lymphoma.
10.**(A).** High-risk HPVs include types 16 and 18 but also types 31, 33, 35, 45, and 56, which are also commonly found in women without lesions. The low-risk HPVs, including 6 and 11 that are commonly found in genital warts, are almost never associated with malignancies.
11.**(A).**
12.**(E).**
13.**(C).** More favorable characteristics include a rapid response to therapy, hyperdiploidy, trisomy of specific chromosomes (4, 10, and 17), and rearrangements of the ETV6-RUNX1 (formerly TEL-AML1) genes.
14.**(B).** Infants with ALL, along with patients who present with specific chromosomal abnormalities, such as t(4; 11), have an even higher risk of relapse despite intensive therapy. However, the poor outcome of Philadelphia chromosome positive ALL with

t(9;22) has dramatically changed by the addition of imatinib to an intensive chemotherapy backbone.

15**.(E).**

16**.(C).** Pharmacogenetic testing of the thiopurine S-methyltransferase (TPMT) gene, which encodes one of the metabolizing enzymes of mercaptopurine, can identify patients who are wild type (normal TPMT enzyme activity), heterozygous (slightly decreased TPMT enzyme activity), or homozygous (low or absent enzyme activity). Decreased TPMT enzyme activity results in an accumulation of a toxic metabolite of mercaptopurine and results in severe myelosuppression, requiring dose reductions of the chemotherapy.

17**.(E).** Testicular relapse occurs in less than 2% of boys with ALL, usually after completion of therapy. Such relapse occurs as painless swelling of one or both testes. The diagnosis is confirmed by biopsy of the affected testis. Treatment includes systemic chemotherapy and possibly local irradiation. A high proportion of boys with a testicular relapse can be successfully retreated, and the survival rate of these patients is good.

18**.(E).**

19**.(B).** Discrete masses, known as chloromas or granulocytic sarcomas, can occur in the absence of apparent bone marrow involvement and typically are associated with a t(8;21) translocation. Chloromas also may be seen in the orbit and epidural space.

20**.(C).** In AML, patients with Down syndrome have much better outcomes than non–Down syndrome children, with a >80% long-term survival rate.

21**.(E).** Approximately 10% of neonates with Down syndrome develop a transient leukemia or myeloproliferative disorder which usually resolves within the 1st 3 mo of life. Although these neonates can require temporary transfusion support, they do not require chemotherapy unless there is evidence of life-threatening complications. However, those patients require close follow-up, because 20-30% will develop typical leukemia by 3 yr of life.

22**.(E).** Twenty-thirty percent of patients who have Down syndrome and who develop transient leukemia or myeloproliferative disorder will develop typical leukemia (often acute megakaryocytic leukemia) by 3 yr of life (mean onset, 16 mo).

23**.(A).** More than 80% of the cases of infantile ALL demonstrate rearrangements of the MLL gene, found at the site of the 11q23 band translocation, the majority of which are the t(4;11). This subset of patients largely accounts for the very high relapse rate.

24.**(A).**

25**.(D).** Stage II includes involvement of 2 or more lymph node regions on the same side of the diaphragm (II) or localized involvement of an extralymphatic organ or site and 1 or more lymph node regions on the same side of the diaphragm (IIE). HL is subclassified into A or B categories: A is used to identify asymptomatic patients and B is for patients who exhibit any B symptoms.

26**.(D).** The primary site of tumor involvement and metastasis pattern varies by pathologic subtype. Lymphoblastic lymphoma (LBL) commonly manifests as an intrathoracic or mediastinal supradiaphragmatic mass and also has a predilection for

spreading to the bone marrow and CNS. Burkitt lymphoma (BL) commonly manifests as abdominal (sporadic type) or head and neck (endemic type) tumor and can metastasize to the bone marrow or CNS. Diffuse large B-cell lymphoma (DLBCL) commonly manifests as either an abdominal or mediastinal primary and, rarely, disseminates to the bone marrow or CNS. ALCL manifests either as a primary cutaneous manifestation (10%) or as systemic disease (90%) with dissemination to liver, spleen, lung, or mediastinum. Bone marrow or CNS disease is rare in ALCL.

27.**(D).** TLS can occur from rapid cell turnover, which is especially common in Burkitt lymphoma.

28**.(B).**

29.**(D).** Recombinant urate oxidase is preferred in patients with a high risk of tumor lysis. Frequently, only a single dose is needed; however, repeat doses can be given if a subsequent rise in uric acid is seen.

30**.(C).** With complete surgical resection the overall survival approaches 80-100%.

31**.(C).** Myxopapillary ependymoma (WHO grade I) is a slow-growing tumor arising from the filum terminale and conus medullaris; it carries good prognosis.

32.**(B).**

33**.(A).** Craniospinal radiation in children younger than 3 yr of age results in severe late neurologic sequelae, including microcephaly, learning disabilities, cognitive impairment, and neuroendocrine dysfunction. Similarly, in older children, late sequelae, such as learning disabilities, neuroendocrine dysfunction, and/or second malignancies, can occur.

34**.(C).** There is no role for chemotherapy in craniopharyngioma.

35**.(D).** Surgical resection is the primary treatment approach for focal and dorsally exophytic tumors and leads to a favorable outcome. Histologically, these 2 groups usually are low-grade gliomas. Cervicomedullary tumors, owing to their location, may not be amenable to surgical resection but are sensitive to radiation therapy. Diffuse intrinsic tumors, characterized by the diffuse infiltrating pontine glioma, are associated with a very poor outcome independent of histologic diagnosis.

36**.(A).**

37**.(D).** Ventriculoperitoneal shunts have been known to allow extraneural metastases, primarily within the peritoneal cavity but also systemically.

38**.(C).**

39**.(B).** Hyperdiploidy confers better prognosis if the child is younger than 1 yr of age at diagnosis. Amplification of MYCN is strongly associated with advanced tumor stage and poor outcomes. Other chromosomal abnormalities, including loss of heterozygosity of 1p, 11q, and 14q, and gain of 17q, are commonly found in neuroblastoma tumors and are also associated with worse outcomes.

40**.(A).** Horner syndrome, characterized by unilateral ptosis, myosis, and anhidrosis, is associated with a thoracic or cervical primary tumor (e.g., superior cervical ganglion). Symptoms do not resolve with tumor resection.

41**. (C).** The most common sites of metastasis are the regional or distant lymph nodes, long bones and skull, bone marrow, liver, and skin. Lung and brain metastases are rare, occurring in >3% of cases.
42**.(D).** Horner syndrome results from direct invasion of the neural foramina by tumor, causing nerve root compression. opsoclonus– myoclonus–ataxia syndrome is of paraneoplastic nature of autoimmune origin. Neuroblastoma can produce catecholamines that can cause increased sweating and hypertension; it may release vasoactive intestinal peptide, causing a profound secretory diarrhea.
43**.(E).** Infants younger than 1 yr of age also can present in unique fashion, termed stage 4S, with widespread subcutaneous tumor nodules, massive liver involvement, limited bone marrow disease, and a small primary tumor without bone involvement or other metastases.
44**.(E).** The infant has stage 2A and is stratified as low risk. The usual treatment for children with low-risk neuroblastoma is surgery for stages 1 and 2 and observation for stage 4S with cure rates generally >90% without further therapy.
45**.(D).** High-dose chemotherapy with autologous stem cell rescue, and not allogenic bone marrow transplantation, is one of the treatment modality for high risk patients.
46**.(D).** Although biopsy is a reliable diagnostic tool, it is discouraged as it results in disease upstaging. A core needle biopsy obtained via a posterior approach should be performed in cases of unusual presentation (older age, signs of infection, inflammation) or unusual imaging findings (significant adenopathy, no renal parenchyma seen, intratumoral calcification).
47**.(A).** Older age, more than 5 years, is an indication to perform renal biopsy.
48**.(C).** The child is stratified as stage III; so he needs chemo-radiotherapy.
49**.(A).** Prognostic factors for risk-adapted therapy include age, stage, tumor weight, and loss of heterozygosity at chromosomes 1p and 16q. Histology plays a major role in risk stratification of WT. Absence of anaplasia is considered a favorable histologic finding.
50**.(C).** Factors associated with a favorable outcome after relapse include low stage (I/II) at diagnosis, treatment with vincristine and actinomycin D only, no prior radiotherapy, favorable histology, relapse to lung only, and interval from nephrectomy to relapse 12 mo or longer.
51**.(C).** Mesoblastic nephroma is the most common solid renal tumor identified in the neonatal period. Most of the patients are diagnosed before 3 mo of age, whereas Wilms` tumor is rarely diagnosed before 6 mo of age. Radical nephrectomy is the treatment of choice and may be sufficient by itself.
52**.(C).** Bone is the most common site of distant metastasis in clear cell sarcoma of the kidney followed by lung, abdomen, retroperitoneum, brain, and liver.
53**.(E).** Unlike the case for adult RCC, local lymph node involvement is not a poor prognostic indicator in pediatric RCC. Nephrectomy alone may be adequate for early-stage RCC.
54**.(C).** Alveolar tumors occur most often in the trunk and extremities.

55**.(E).** Favorable sites include female genital, paratesticular, and head and neck (nonparameningeal) regions; all other sites are considered unfavorable. Involvement of paranasal sinuses by rhabdomyosarcoma is considered as unfavorable site.
56**.(D).** Parosteal osteosarcoma is a low-grade, well-differentiated tumor that does not invade the medullary cavity and most commonly is found in the posterior aspect of the distal femur. Surgical resection alone often is curative in this lesion, which has a low propensity for metastatic spread.
57**.(C).** All the mentioned factors carry poor prognosis, but multiple bony metastases at the time of diagnosis is the worst.
58**.(D).** Osteosarcoma is a radio-resistant tumor.
59**.(C).** Osteoid osteomas are diagnosed between 5 and 20 yr of age. The clinical pattern is characteristic, consisting of unremitting and gradually increasing pain that often is worst at night and is relieved by aspirin. Boys are more affected than girls. Radiographs are distinctive, showing a round or oval metaphyseal or diaphyseal lucency (nidus) surrounded by sclerotic bone.
60**.(B).** Imaging studies are not diagnostic, and biopsies are contraindicated. Indirect ophthalmoscopy with slit-lamp evaluation can detect retinoblastoma tumors, but a complete evaluation requires an examination under general anesthesia by an experienced ophthalmologist to obtain complete visualization of both eyes, which also facilitates photographing and mapping of the tumors.
61**.(B).** Retinoblastoma can be either hereditary or sporadic. Hereditary cases usually are diagnosed at a younger age and are multifocal and bilateral, whereas sporadic cases are usually diagnosed in older children who tend to have unilateral, unifocal involvement. All 1st-degree relatives of children with known or suspected hereditary retinoblastoma should have retinal examinations to identify retinomas or retinal scars.
62**.(B).** Infants normally have higher levels of AFP, which fall to normal adult levels by about age 8 mo; consequently, high AFP levels must be interpreted with caution in this age group.
63**.(C).** Elevation of the β subunit of human chorionic gonadotropin, which is secreted by syncytiotrophoblasts, is seen with choriocarcinoma and germinomas.
64**.(C).** Cisplatin-based chemotherapy regimens usually are curative in GCTs that cannot be complete5ly resected, even if metastases are present.
65**.(E).** Hepatitis B or C infection is often associated with hepatocellular carcinoma.
66**.(A).** The pure fetal histology subtype predicts a more favorable outcome and the small cell undifferentiated subtype is associated with normal AFP levels and predicts a worse outcome.
67**.(C).** The risk of hemangioma is 3-5 times higher in girls than boys. The risk is doubled in premature infants and 10 times higher in offspring of women who had chorionic villus sampling.
68**.(D).** An ultrasonographic scan or MRI of the liver should be performed if multiple cutaneous lesions are present.

69**.(E).** All the features of Kasabach-Merritt syndrome results from platelet and red blood cell trapping and activation of the clotting system within the vasculature of the hemangioma. This syndrome is associated with kaposiform hemangioendotheliomas or tufted angiomas but not with infantile hemangiomas.
70**.(C).** Localized cystic hygroma lesions may be surgically resected, but this can be difficult, owing to their infiltrative nature. Recurrence is common with incompletely resected lesions. Aspiration can provide temporary relief in an emergency and reaccumulation will occur. Treatment by injection of sclerosing agents (by streptococcal immunotherapeutic agent OK-432) is the treatment of choice; its use will prevent the need for surgery in most cases. Laser therapy is not used in such age. Although spontaneous regression has been reported but is not typical.
71**.(D).** PTC has an excellent overall prognosis even in the presence of metastatic disease. The primary therapy is a total thyroidectomy and lymph node dissection. 131I is used postoperatively to treat distant metastasis and unresectable residual neck disease. The TSH level is suppressed by giving supraphysiologic levothyroxine as TSH stimulates tumor growth. Long-term follow-up involves monitoring of thyroglobulin tumor markers.
72**.(E).** Spitz nevus is a benign harmless skin lesion. However, making the distinction from diagnosis of melanoma can be difficult.
73**.(B).** Patients with fair skin are at particularly high risk for development of melanoma.
74**.(E).** Most pediatric patients present with advanced locoregional disease. LDH is elevated but it is not specific. Epstein-Barr virus DNA levels correlate with disease stage, have prognostic valueand can be used to monitor for recurrence.The outcome depends on the extent of disease; patients with distant metastases have a very poor prognosis.
75**.(A).** LCH has an extremely variable presentation. The skeleton is involved in 80% of patients and may be the only affected site, especially in children older than 5 yr of age. Bone lesions may be single or multiple and are seen most commonly in the skull**.**
76**.(D).** Bone is the most common site of involvement by LCH. About 50% of patients experience skin involvement at some time during the course of disease. Localized or disseminated lymphadenopathy is present in approximately 33% of patients. Otitis media is present in 30-40% of patients. In 10-15% of patients, pulmonary infiltrates are found on radiography. The lesions may range from diffuse fibrosis and disseminated nodular infiltrates to diffuse cystic changes.
77**.(A).**